

Case Report

Papillary thyroid carcinoma in a thyroglossal duct cyst: A case report of a 40-year-old female patient at Hawassa University Comprehensive Specialized Hospital

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Abstract

Thyroglossal duct cyst (TDC) is a common congenital anomaly of the thyroid gland, usually presenting as a midline neck mass. The presence of malignancy occurring within these cysts is a rare condition, accounting only for 1% of all cases of thyroglossal duct cyst. Usually, the diagnosis is only made postoperatively after excision of the cyst. Given the rarity of thyroglossal duct cyst carcinoma, definitive treatment is not well agreed upon. The Sistrunk's procedure can be the sole procedure or it can be followed by total thyroidectomy, nodal dissection, and postoperative radioactive iodine treatment. Here, we present a case of papillary carcinoma of the thyroglossal duct cyst in a 40-year-old female patient who presented with a painless anterior neck swelling above the thyroid cartilage. She had an ultrasound guided FNAC of the mass which suggested papillary thyroid carcinoma. She then underwent a Sistrunk's procedure and has had an uneventful post-operative course. Postoperative histopathology showed papillary thyroid carcinoma with surrounding fibro fatty tissue remnant. Interestingly, no normal thyroid tissue was noted in the specimen. Papillary carcinoma of TDC, which is the most common carcinoma of TDC, should only be treated with the Sistrunk's procedure and strict long-term follow up, in the absence of high-risk features. The purpose of this case report is to increase the awareness among physicians on how to approach this rare disease, including its work up and entertain the various management options in addition to the Sistrunk's procedure.

Key words: Thyroglossal duct cyst; Papillary thyroid carcinoma; Sistrunk's procedure; FNAC

Introduction

During the embryologic development of the thyroid gland, the thyroglossal tract involutes between 7-10 weeks of gestation (1,2). A part of the thyroglossal tract may fail to obliterate and become a thyroglossal duct cyst (3). A thyroglossal duct cyst (TDC) is a very common anomaly in the anterior neck (4,5). It is twice as common as branchial cleft cysts (2). Nearly 70% of anterior neck masses in children and about 7% of anterior neck masses in adults is due to a thyroglossal duct cyst (3,4,6,7). Thyroglossal duct cysts commonly complicate with an infection and sinus tract formation but thyroid carcinoma is a rare complication of thyroglossal duct cysts occurring in less than 1 % of cysts (8 - 10). Carcinoma should be considered if the mass is hard, fixed irregular or there is associated palpable cervical lymphadenopathy (11,12). "The first case of thyroglossal duct cyst carcinoma was reported in 1911." (6,13) Currently only 275 cases are reported in the literature (6).

"Generally there are two theories to explain the thyrogenic origin of TDC adenocarcinomas. Firstly, the de novo theory is based on the fact that in 62% of cases, ectopic thyroid tissue can be identified histopathologically, and this is supported by the absence of a medullary carcinoma in the TDC as it arises from Para follicular cells. The second is the metastatic theory which suggests that TDC carcinoma is metastatic from an occult primary thyroid gland, as papillary carcinoma is multifocal in nature. As a result, there is no consensus as to whether total thyroidectomy should be performed immediately after initial diagnosis of a carcinoma arising in a TDC."(5,8,11)

Papillary carcinoma is the most common histologic variant occurring in thyroglossal duct cysts. It accounts for 75-80% of thyroglossal duct cyst carcinomas (4,11,13,14). The other histologic variants include Follicular carcinoma

and mixed types. Squamous cell carcinoma is an even rarer histologic variant arising with in TDC and it accounts for less than 5% of the cases (5,6,8,11). As it is the case with thyroglossal duct cysts, carcinomas of these cysts present as an asymptomatic anterior midline neck mass (4). Diagnosis of TDC carcinoma cannot be made on clinical examination, neck ultrasound and at FNAC. It is often made after surgery (7,8). The majority of carcinomas are confined to the cyst (4). Local invasiveness was noted in about 1-4% of TDC carcinomas. Cervical nodal metastasis occurs in about 7%-25% of cases and distant metastasis occurs in around 1.3% of cases which is very low compared to carcinomas arising from the thyroid gland (11,13). The average age of presentation is 40yr and women are commonly affected (7,9).

There are no well-set treatment guidelines for carcinomas of thyroglossal duct cysts given that the disease is very rare (1). Carcinomas confined to the thyroglossal duct cyst are treated with Sistrunk's procedure and regular follow up, where as the more aggressive ones' require total thyroidectomy, radioiodine ablation and thyroid stimulating hormone suppression in addition to the Sistrunk's procedure (3,5,13).

The purpose of this case report is to increase the awareness among physicians on how to approach this rare disease, including its work up and entertain the various management options in addition to the Sistrunk's procedure.

Case description

We present a case of a 40-year-old female patient who presented to Hawassa University Comprehensive Specialized Hospital with a gradually increasing painless anterior neck swelling above the level of the thyroid gland for 2 months. Her medical history was

unremarkable. No clinical features of thyroid dysfunction were noted.

There was no history of dysphagia, dyspnea, or hoarseness. The patient had not been previously exposed to radiation or other known carcinogens. Familial medical history was negative for thyroid gland or neoplastic diseases. On physical examination, she had normal body temperature, pulse rate of 78 beats/min and was normotensive.

Lymphoglandular examination revealed a firm, non-tender, a 4x2cm well-demarcated mass in the upper anterior neck above the level of the thyroid cartilage and over the midline (see Fig. 1)



Fig. 1: The patient on the OR table

The mass was noted to move with deglutition and tongue protrusion. The overlying skin demonstrated no sign of inflammation. Both lobes of the thyroid gland were not palpable. No palpable cervical lymph node was appreciated. The remainder of her systemic examination was normal.

On biochemical analysis, her free T3, free T4 and TSH level were found within normal ranges- 5.2 pmol/L (normal range: 2.8-9.5 pmol/L), 22.7 pmol/L (normal range: 9.5- 25.5 pmol/L) and 0.477 mIU/L (normal range: 0.3-5.0 mIU/L), respectively. At entry serum chemistry tests, electrocardiogram and chest X-rays were normal.

A neck ultrasound identified a 4 cm cyst above a normal-appearing thyroid gland. The ultrasound guided FNAC of the midline mass was suggestive of papillary thyroid carcinoma.

A Sistrunk's surgical procedure was then performed and the thyroglossal duct was removed, including the entire duct from the gland to the level of the foramen cecum and the middle portion of the hyoid bone (see Fig. 2). There were no enlarged nodes on level 1 and 2 LNs. The excisional biopsy showed all morphologic features of papillary thyroid carcinoma with surrounding fiber-fatty tissue remnant. Normal thyroid parenchyma was not represented (see Fig. 3).



Fig. 2: Intraoperative photograph while doing Sistrunk's Procedure

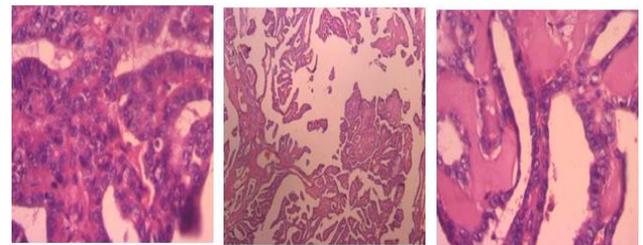


Fig. 3: Section shows capsulated tissue fragment contacting papillary architecture lined by oval cells exhibiting nuclear grooving, clearing and pseudo-inclusions. The adjacent area of bland colloid filled follicles are also noted.

Post-operative follow-up was uneventful and the patient left the hospital on her 3rd postoperative day.

Discussion

Normally the embryonic route of the developing thyroid gland gets obliterated and disappears by the 10th week of development.(4,5,13) “A thyroglossal remnant can remain in the form of a cyst, a tract or duct, or as ectopic thyroid within a cyst or duct.”(4,12) The two common complications of a thyroglossal duct cyst are infection and sinus formation. Yet, development of carcinoma within the duct is a rare phenomenon (4). TDC Carcinoma commonly affects women than men with female: male ratio of 2.1:1. Patients with TDC commonly present with an average age of 40 yrs (1,11). This was the case with our patient who is a 40 year old female patient. All cases of TDC carcinomas should be investigated preoperatively with neck ultrasound, CT scan, MRI and FNA (6). Even after doing all these investigations, definitive diagnosis is usually made postoperatively with histopathologic examination. Our patient had neck Ultrasound and Ultrasound guided FNAC of the mass. Normally the yield of FNAC increases when it is Ultrasound guided (2,6). Certain criteria have to be met to make diagnosis of primary TDC carcinomas like: cancer being present within the wall of TDC and absence of cancer in another primary site. In 11%-40% cases of TDC carcinomas there can be concomitant thyroid carcinomas (9).

There is a lot of controversy with regard to the definitive treatment of TDC carcinoma (4). The initial step in the management is straight forward which is to do the Sistrunk's procedure; the controversy lies whether additional procedures are needed or not, like Total thyroidectomy, radioactive iodine and thyroid suppression (9,12). For example, total thyroidectomy can be associated with several morbidities like: hypocalcaemia and recurrent laryngeal nerve

injury and the need for lifelong thyroid hormone replacement; hence the decision of doing it is not easy (9).

Sistrunk's procedure is said to have a better survival than simple mass excision; Hence, it should be performed in all cases. This procedure consists of removal of the thyroglossal duct cyst, the central portion of the hyoid bone, and the thyroglossal duct all in continuity (6,13). This procedure has a recurrence rate of less than 4% and may complicate in about 1% of the cases (2,13). Patients with labeled high characteristics are candidates to undergo Sistrunk's procedure plus total thyroidectomy (1). “High-risk characteristics that have been proposed by some include age over 45yrs, tumors with a diameter greater than 1.5cm, previous radiation exposure, presence of nodal disease, invasion of cyst wall, positive histopathologic margins, and presence of a tumor in the thyroid on radiological evaluation.”(1,11,13) Fortunately, only 1-4% of TDC carcinomas are locally invasive and not more than 11% of cases are associated with cervical lymph node metastasis (13). In fact, some authors recommend adding total thyroidectomy has little significance on recurrence and survival (13). In our patient, due to lack of high risk features the patient has undergone only the Sistrunk's procedure (1). Elective neck dissection is not generally recommended in all cases of TDC carcinomas as the risk of metastasis is very low. So, it should be done in cases of clinically positive nodes (3).

”The prognosis of patients with TDC carcinoma is usually very good. In a review by Patel et al., with a median follow-up of 71months, the 5-year and 10-year Kaplan-Meier overall survival (OS) was 100% and 95.6%, respectively.”(4,11) It is the worst with squamous cell carcinoma (5).

It has been reported that the cure rate of Sistrunk's procedure for papillary thyroid carcinoma of TDC reaches up to 95% (11). “Patel et al in 2002 showed that univariate

analysis of prognostic factors predictive of overall survival revealed that the only significant predictor of outcome was the extent of surgery for TDC. The addition of total thyroidectomy didn't have a significant impact on outcome ($p=0.1$).”(10)

Careful long-term follow up, whether total thyroidectomy is done or not, is very essential to pick any recurrences of papillary carcinoma of TDC (11). Hence, the follow up consists of physical examination and neck scan every six month for the first year then annually afterwards (4,13). Our case was seen on the 2nd month of her postoperative day (see Fig. 4) and she is currently on her 1 year and 2 months post-operative course and she has no subjective complaint, no neck swelling and no abnormality detected on neck ultrasound.



Fig .4: The patient on 2nd month's post-operative day

Conclusion

Thyroglossal duct carcinoma is a rare entity which has to be considered in patients presenting with midline anterior neck masses. Histopathology may allow a determination of whether this malignancy in the thyroglossal duct cyst is primary or due to a secondary metastasis from a thyroid malignancy-information which is useful in recognizing prognosis as well as for formulating treatment. Papillary carcinoma of

TDC, which is the most common carcinoma of TDC, should only be treated with the Sistrunk's procedure and strict long-term follow up. This was the case in our patient. But in the presence of high risk features, in addition to Sistrunk's procedure patients should undergo total thyroidectomy and radioiodine ablation.

Acknowledgement

The authors would like to express sincere gratitude to colleagues in the American College of Surgeons for their encouragement in undertaking this case report. We would also like to thank the Department of Pathology for providing us with pathologic slides. The paper would not have been realized without the immense cooperation of the patient in taking detailed history, doing appropriate physical examination, and taking pictures when needed.

Consent for publication

The patient consented for the publication of the present case report.

Data Availability statement

Not applicable.

Conflict of interest

The authors declare that they have no conflicts of interest.

Authors' Contribution

ABA is the PI who took part in the entire process from the inception of the case report to its final write up. In addition he took part in the operation

of the case. TTT took part in the operation and reviewed the initial draft of the case report. ATL took part in the operation and provided the patient's history and investigation results. KLL took part in the reviewing of the initial draft of the case report. All authors approved the final manuscript.

References

1. Van Beck J, Khaja SF. Thyroglossal Duct Cyst Carcinoma in a Young Female: Case Report and Review of Literature. *Case Rep Otolaryngol*. 2019;2019(Figure 1):1–3.
2. Maleki N, Alamdari M, Feizi I, Tavosi Z. Papillary thyroid carcinoma arising from a thyroglossal duct cyst. *Arch Int Surg*. 2013;3(3):229.
3. Balalaa N, Megahed M, Al Ashari M, Branicki F. Thyroglossal duct cyst papillary carcinoma. *Case Rep Oncol*. 2011;4(1):39–43.
4. Alatsakis M, Drogouti M, Tsompanidou C, Katsourakis A, Chatzis I. Invasive thyroglossal duct cyst papillary carcinoma: A case report and review of the literature. *Am J Case Rep*. 2018;19:757–62.
5. Gupta N, Dass A, Bhutani M, Singhal SK, Verma H, Punia RPS. Papillary carcinoma in thyroglossal duct cyst: An unusual case. *Egypt J Ear, Nose, Throat Allied Sci* [Internet]. 2014;15(1):45–7. Available from: <http://dx.doi.org/10.1016/j.ejenta.2013.10.001>.
6. Verma R, Patro SK, Damodharan N, Sood A, Bal A. Papillary carcinoma thyroid in a thyroglossal cyst: A management dilemma. *Acta Oto-Laryngologica Case Reports* [Internet]. 2017;2(1):5–10. Available from <https://doi.org/10.1080/23772484.2016.1270166>
7. Penna GCE, Mendes HG, Kraft AO, Berenstein CK, Fonseca B, Martorina WJ, et al. Simultaneous Papillary Carcinoma in Thyroglossal Duct Cyst and Thyroid. *Case Rep Endocrinol*. 2017;2017.
8. Chrisoulidou A, Iliadou PK, Doumala E, Mathiopolou L, Boudina M, Alevizaki M, et al. Thyroglossal duct cyst carcinomas: Is there a need for thyroidectomy? *Hormones*. 2013;12(4):522–8.
9. Saavedra-Leveau JL, Chang-Grozo SL, Dominguez-Prado ME, Ticona-Zegarra LA. “Incidental papillary thyroid cancer in thyroglossal duct cyst”: A case report. *Iran J Otorhinolaryngol*. 2021;33(3):187–90.
10. Manipadam JM, Manipadam MT, Thomas EM, Michael RC, Ramakant P, Abraham DT, et al. Thyroglossal duct carcinoma: A case series and approach to management. *World J Endocr Surg*. 2011;3(2):59–63.
11. Ubayasiri KM, Brocklehurst J, Judd O, Beasley N. A decade of experience of thyroglossal cyst excision. *Ann R Coll Surg Engl*. 2013;95(4):263–5.
12. Kandogan T, Erkan N, Vardar E. Papillary carcinoma arising in a thyroglossal duct cyst with associated microcarcinoma of the thyroid and without cervical lymph node metastasis: A case report. *J Med Case Rep*. 2008;2:1–3.
13. Arslan I, Eruyar A, Gumussoy M, Genc S. Conservatively managed thyroglossal duct cyst carcinoma with lymph node metastasis: Long-term result. *Thyroid Res Pract*. 2016;13(2):77.
14. Ziółkowska M, Bień S, Sygut J, Klimas A, Zylka S. Papillary thyroid carcinoma in thyroglossal duct cyst. *Otolaryngol Pol*. 2009;63(1):50–3.